

**CASE REPORT**

Volume 7 Issue 3

***Plasmablastic Lymphoma of the Kidney in a Previously Undiagnosed HIV Patient*****Amrita Valluri, BS<sup>1</sup>, Adarsh Sidda, MD<sup>1</sup>, Anisha Valluri, BS<sup>1</sup>, Gurusidda Manu, MD<sup>2</sup>, Niru Nahar, MD<sup>1</sup>****ABSTRACT**

We present the case of a 61-year-old male who presented with bilateral flank pain and dysuria. A biopsy and histopathological analysis of a mass found in the left kidney led to the diagnosis of plasmablastic lymphoma (PBL). Initial suspicion of PBL was low, as the kidney is not a typically involved organ.<sup>1,2</sup> As a part of routine workup, HIV testing was ordered resulting in positive preliminary and confirmatory results. The patient denied awareness of his HIV status. Chemotherapy with uricosuric agents and antiretroviral therapy was initiated, but the patient developed tumor lysis syndrome with phosphate nephropathy leading to discontinuation of the regimen. Due to deterioration in clinical status, the patient chose palliative measures. This case report addresses the diagnostic challenges of plasmablastic lymphoma and its rare presentation in an unusual location.

**KEYWORDS**

Plasmablastic lymphoma, Immunocompromised, Atypical location

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**INTRODUCTION**

Plasmablastic lymphoma (PBL) is a rare variant of non-Hodgkin lymphoma. Characteristic findings are a lack of pan B-cell markers, high Ki-67 index, EBV positivity, and MYC gene rearrangement.<sup>1</sup> Around 65% of cases of PBL arise in immunocompromised individuals.<sup>1</sup> Despite its higher prevalence in this population, only a small minority of patients present with PBL as the initial symptom of immunocompromised status.<sup>1,2,3</sup> We present the case of a 61-year-old male diagnosed with plasmablastic lymphoma of the kidney who later tested HIV-positive. Prior to diagnosis, clinical suspicion of PBL was low as the kidney is not a commonly reported site of presentation in the current literature.<sup>1,2</sup>

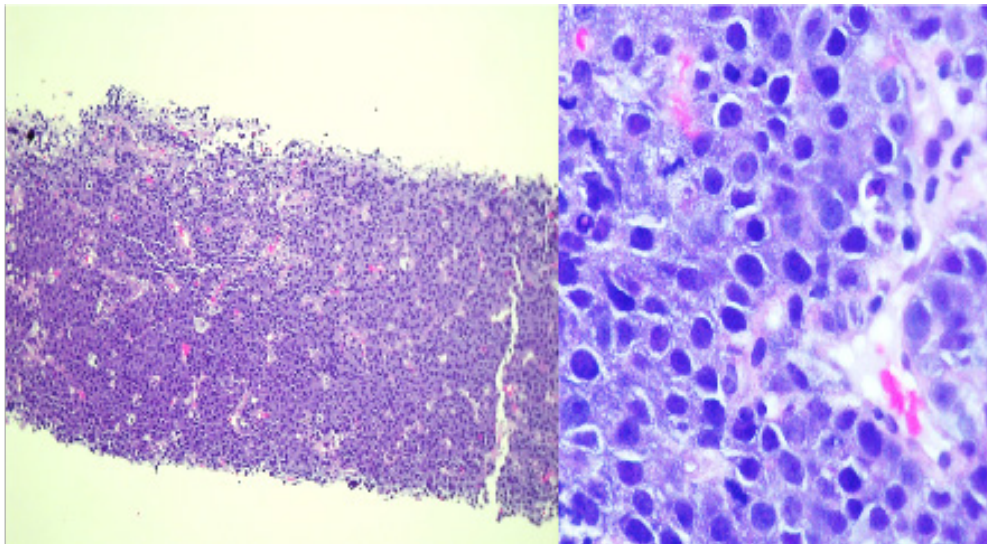
**CASE PRESENTATION**

Our patient is a 61-year-old male with a past medical history of hypertension and blindness who presented with bilateral flank pain and dysuria. Initial laboratory indices showed acute kidney injury with creatinine of 1.61 and a lactic acid level of 8.82. CT of the abdomen showed a large mass in the left kidney extending along the ureter inferiorly towards the bladder.

A specimen obtained through CT-guided biopsy consisted of diffuse sheets of large plasmablastic cells with abundant cytoplasm, vesicular chromatin, and prominent centrally located nucleoli (Figures 1A and 1B). Frequent mitotic figures, apoptotic bodies, and intermixed tingible body macrophages were noted. Other pertinent findings included neoplastic cells positive for CD45, CD138, CD56, BCL-2, CD10, MYC, MUM1, and scattered BCL-6. Immunostains for PAX-5, CD79a, and EMA were patchy positive with Ki-67 index > 90 with kappa-predominant cells present. CD20, CD3, cyclin D1, CD34, CD30, ALK-1, CD4, synaptophysin, chromogranin, GATA-3, CK7, and CK20 immunostains were negative. Several notable findings including lack of pan B-cell markers, EBV positive RNA, MYC gene rearrangement, and >90% Ki-67 immunostains led to the diagnosis of PBL (Figures 2A, 2B, 2C).

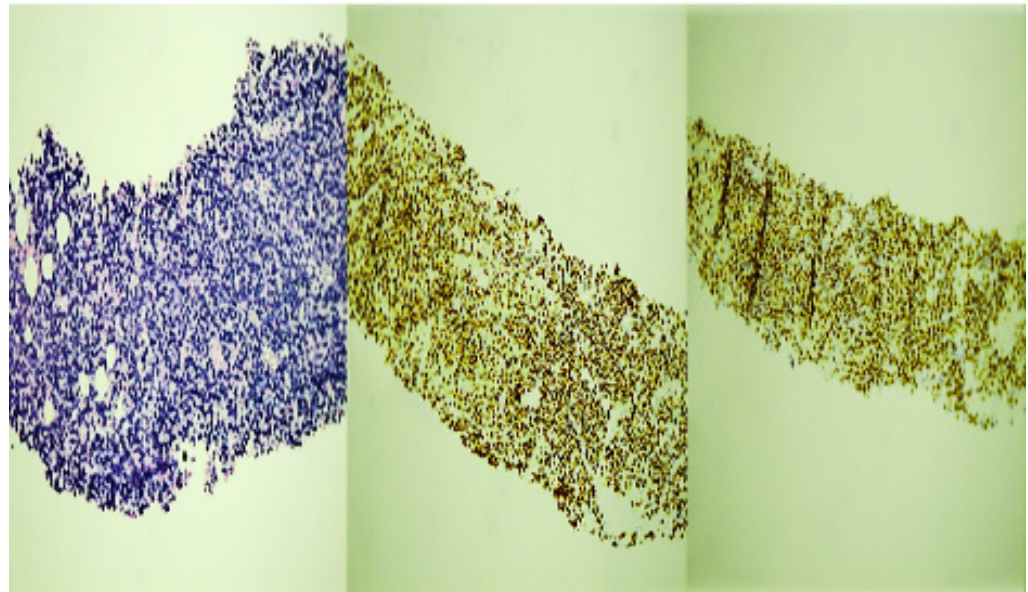
After the diagnosis of PBL, a subsequent PET scan visualized the mass in the left kidney with extensive lymph node, bone, liver, and muscle metastasis. As part of routine workup, HIV testing was ordered. Initial and confirmatory HIV tests were positive, although the patient denied any awareness of infection. He denied IV drug use, blood transfusions, or a





**FIGURE 1.** (H & E stain): 1A: Low power showing diffuse proliferation of monomorphic large cells; 1B: High-power showing large cells with plasmablastic features.

history of other STDs. Upon further testing, the patient's CD4 count was found to be 81, and he was given a diagnosis of AIDS. The patient was started on infection prophylaxis and antiretroviral therapy (ART) consisting of tenofovir/emtricitabine and dolutegravir. A chemotherapy regimen of Etoposide, Prednisone, Vincristine, Cyclophosphamide, and Doxorubicin (DA-EPOCH) was initiated with Allopurinol and a dose of Rasburicase. During his first cycle of chemotherapy, the patient developed tumor lysis syndrome with phosphate nephropathy. His clinical status deteriorated with worsening respiratory status. At this time, the patient decided against pursuing aggressive treatment measures. His code status was changed to comfort measures upon request.



**FIGURE 2.** (Immunohistochemical stains and EBER): 2A: Positive for In Situ Hybridization of EBV-encoded RNA; 2B: high Ki-67 proliferation index; 2C: Positive staining C-Myc.

## DISCUSSION

Plasmablastic lymphoma commonly occurs in the setting of immunodeficiency.<sup>4</sup> With regards to HIV infection, the incidence of PBL is approximately 2% of all HIV-related lymphomas.<sup>5</sup> The differential diagnosis in our patient included plasmablastic multiple myeloma, Burkitt's lymphoma, EBV positive diffuse large



B-cell lymphoma, and extra cavitory primary effusion lymphoma. These pathologies were ruled out due to a lack of reconciliation with the histopathological features identified in the tumor sample. There are characteristics unique to PBL that help distinguish it from other etiologies. Biopsied samples lack pan B-cell lymphoma markers, and Ki-67 staining index is typically high.<sup>7</sup> Characteristic immunophenotype markers seen in PBL include CD138, CD38, CD79, and MUM1.<sup>6</sup> Expression of CD4, CD10, and CD56 have also been reported.<sup>6</sup> MYC gene rearrangement is found in the majority of PBLs, thus raising suspicion if present.<sup>8</sup> In HIV-positive patients, EBV RNA is commonly detected.<sup>9</sup> Our patient expressed CD138, CD56, BCL-2, CD10, MUM1, and scattered BCL-6 positivity. CD56 and CD10 were positive in the majority of cells. Immunostains for PAX-5, CD79a, and EMA were also patchy positive. The most noteworthy findings our patient had that led to diagnosis were EBV positivity, Ki-67 >90%, and MYC gene rearrangement. Although typical immunohistopathological findings for PBL were present, there are unique features in our patient's case. The oral cavity, gastrointestinal tract, lymph nodes, and skin are the sites with the highest incidence of PBL.<sup>1,2</sup> The location of the tumor within the left kidney found in our patient is an atypical presentation.

Once diagnosed, treatment of PBL comes with challenges. Intensive regimens such as etoposide, vincristine doxorubicin, cyclophosphamide, and prednisone (EPOCH) are preferred over the standard regimens of cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP) used regularly for other non-Hodgkin lymphomas.<sup>10</sup> Other novel therapies such as EBV-targeted cellular immunotherapy, CAR-T cell therapy, and MYC targeting agents are currently being studied in hopes of a better prognostic outcome for this highly aggressive tumor.<sup>11</sup>

## CONCLUSION

Our patient's case suggests that suspicion of PBL should be maintained if a tumor specimen has immunoblastic or plasmablastic markers even if the clinical scenario does not fit the classic presentation. Our case may also support testing for etiologies of immunocompromised status in patients diagnosed with PBL. Though the definitive diagnosis is oftentimes challenging, collaboration with a multidis-

ciplinary team consisting of oncology, pathology, radiology, and infectious disease should be undertaken.

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